Repair of Scimitar Syndrome

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Scimitar syndrome is a complex constellation of vascular, bronchial, and parenchymal malformations of the right lung. This rare congenital malady was first described in autopsy specimens by Cooper[1] in London and Chassinat[2] in Paris in 1836. The hallmark feature is anomalous venous drainage of the right lung that produces a distinctive image on anteroposterior chest roentgenogram (Fig 1) resembling a scimitar, a Turkish sword (Fig 2). It was Neill and associates[3] who recognized this resemblance in 1960 and coined the term "scimitar syndrome" to describe this set of anatomic aberrations.

Typically, the pulmonary (or "scimitar") vein is single, drains the whole lung, courses parallel to the right heart border in a curvilinear path, and terminates in the inferior vena cava below the diaphragm. The classic radiographic appearance is present, in only about one-third of reported cases.[4] In the remaining two-thirds of cases, the vein may be obscured by the right heart border or may be a size and shape that is less prominent radiographically. The vein may be multibranched, which reduces its radiographic density.

The extent of right lung drainage by the anomalous vein also varies. Most typically, scimitar vein drains the entire right lung (Fig 3), but several variations have been reported. The right upper lobe may drain separately into the left atrium while the remainder of the pulmonary venous drainage enters the inferior vena cava. Alternatively, the anomalous vein may drain into the inferior vena cava directly and into the left atrium indirectly via a bridging vein. In another path, the right superior pulmonary vein enters either the right atrium or the superior vena cava and the remainder of the pulmonary venous blood travels to the inferior vena cava via the scimitar vein. There have even been reports of a vein with the typical scimitar appearance that drains the right lung normally into the left atrium. In all cases but the last, the left-to-right shunt via this pulmonary venous return is typically less than 2:1.

Variability in the connection between the anomalous right pulmonary vein and the inferior vena cava has also been described. Typically this connection is subdiaphragmatic.[4,5] However, a supradiaphragmatic venous confluence may also be found.[6]

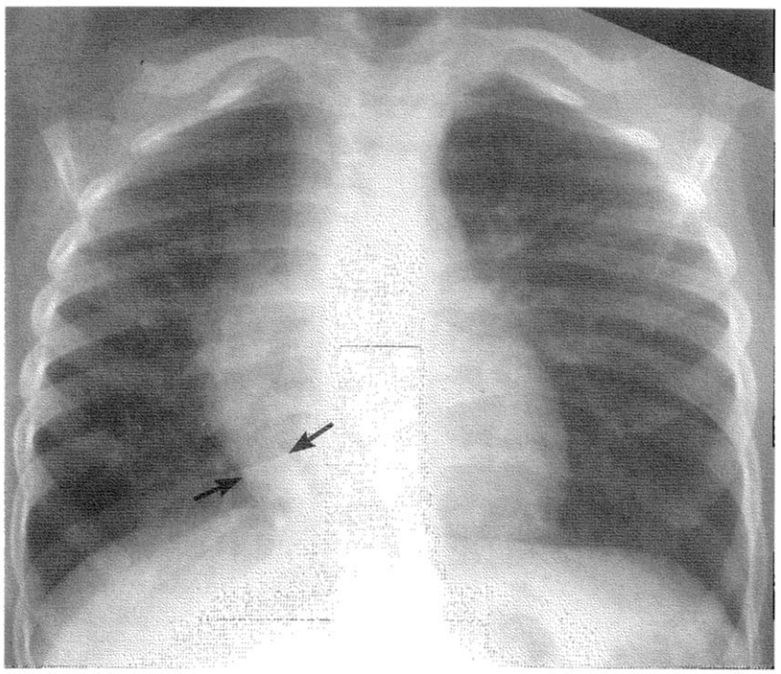
Other malformations in scimitar syndrome may include diminished lung size, bronchial structural and branching defects, and peculiar lobation. Approximately 90% of right lungs are hypoplastic.[4] This diminution in size is likely to be responsible for the dextroversion of the heart present in many cases.

Right bronchial abnormalities have been identified by bronchography, bronchoscopy, or in pathologic specimens and cover a vast array of possibilities. Branching defects occur in both large and small airways. The right upper lobe bronchus may be absent, or the distal airways may be sparsely distributed through the lung parenchyma.[3] In some cases, the branching pattern is the mirror image of that in the left lung. Structurally the bronchi themselves may be small, stenotic, or atretic with areas of diverticulae.[7,8,9]

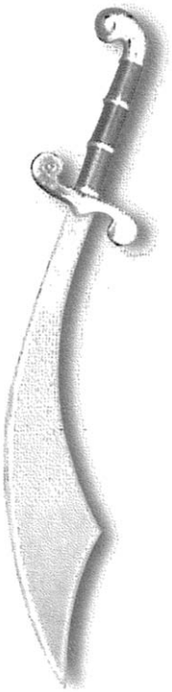
Lobe abnormalities take various forms. Most commonly one lobe is absent, usually the upper lobe, but the middle lobe also may be missing. Hypoplasia of the right lower lobe phenotypically resembles a bilobed lung with a sequestration. But unlike a true sequestration, this hypoplastic lower lobe has direct bronchial communication with the remainder of the bronchial tree.[4] Even a unilobate right lung has been described in a patient with scimitar syndrome.[10]

Arterial blood supply to the malformed right lung is another highly variable component of scimitar syndrome. Sources of blood flow to the right lung include the right pulmonary artery, bronchial arteries, and anomalous systemic arterial collateral vessels that arise principly from the abdominal aorta or its branches. Whereas bronchial artery abnormalities have not been described, abnormalities in the right pulmonary artery are common.

Right pulmonary artery size ranges from normal in 40% of cases[5] to atretic or completely absent in rare cases. Most commonly, this artery is hypoplastic. As might be expected, there is a direct correlation between the size of the right pulmonary artery and the size of its corresponding lung.[4] Massumi and colleagues[11] proposed that pulmonary venous obstruction in the right lung causes venous engorgement and leads to decreased compliance. The relatively noncompliant lung becomes hypoventilated, and alveolar collapse develops. Collapsed alveoli compress capillaries and, coupled with pulmonary venous congestion, shift pulmonary arterial blood flow to the contralateral lung. Consequently, decreased flow in the right pulmonary artery or relatively increased flow to the left lung produces a size discrepancy in the pulmonary arteries proportional to

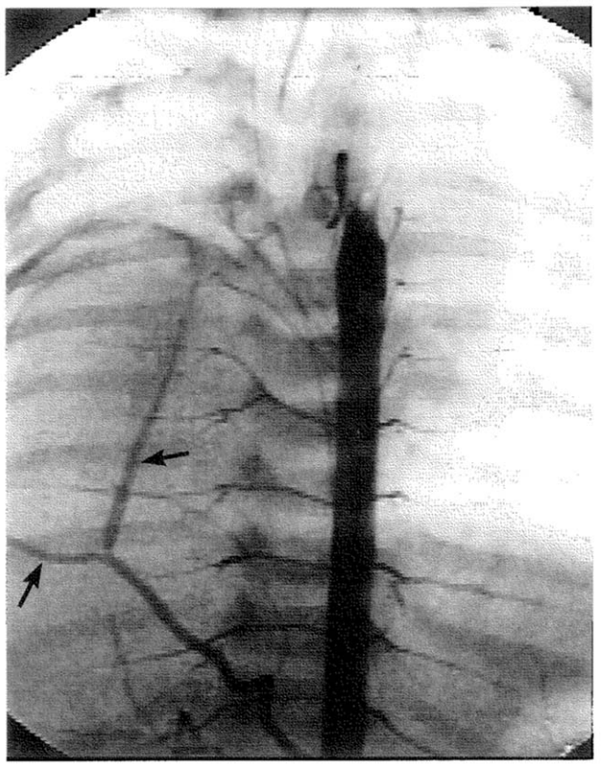


1 Chest radiograph illustrating the appearance and course of the anomalous right pulmonary, or scimitar, vein (arrow).



2 A scimitar.

3 Pulmonary venogram, left anterior oblique projection. The catheter is passed from the groin through the inferior vena cava and directly into the scimitar vein. Contrast injection shows the anomalous vein in its vertical orientation and the acute angle at which it enters the inferior vena cava. Early opacification of the right atrium is evident.



4 Descending aortogram, straight anteroposterior projection. A retrograde catheter is placed in the aorta. Contrast injection shows an aberrant aortopulmonary collateral (arrows) that branches as it approaches the right lung.

flow and to the size of the right lung. Several authors[4,7,11] have suggested that the anomalous systemic arterial collaterals exist to substitute for this diminished blood flow through the pulmonary artery.

The systemic arterial collateral supply to the right lung rarely arises from the thoracic aorta.[4] Most typically, it comes from one or more vessels that originate from the abdominal aorta or its branches (Fig 4). These vessels course obliquely and superiorly to pierce the right leaf of the diaphragm and then travel within the inferior pulmonary ligament to reach the lung surface, where they often branch. Their contribution to perfusion of the lung parenchyma is highly variable, ranging from miniscule in very small vessels to complete systemic supply of the right lung when the right pulmonary artery is absent.[6] Most commonly, however, the systemic arteries supply the right lower lobe or some of its segments.[4,7,8,12] Alternatively, flow from these anomalous arteries may supply both the right lower and middle lobes.[7]

When scimitar syndrome is diagnosed in childhood (particularly in infancy), it is associated with several congenital heart defects with incidence ranging from 24% to 100%.[4,5,8,13] Atrial septal defect is the most common of these, accounting for 20%–83% of defects.[4,5,13-15] Coarctation of the aorta, ventricular septal defect, patent ductus arteriosus, tetralogy of Fallot, and persistent left superior vena cava have also been described.[4,5,13-17] Gao and colleagues[16] reported 7 of 13 infants with left-sided lesions, including hypoplastic left ventricle, arch obstruction, subaortic stenosis, pulmonary vein stenosis, and anomalous left coronary artery, all of which are associated with poor outcome.

Although patients with scimitar syndrome can have various anatomic manifestations of the disorder, all present with either one of two distinct clinical profiles. In the so-called "adult" form, the patient is either asymptomatic or has recurrent respiratory infections, mild exertional dyspnea, heart murmur, or deformation of the right hemithorax. Occasionally, the scimitar sign is detected on routine chest roentgenogram obtained for unrelated reasons. The adult form has been reported in patients age under 1 year to 73 years, and patients generally tolerate the syndrome well.[17-19] These patients typically do not have pulmonary hypertension and have fewer and less severe associated congenital heart defects, such as secundum atrial septal defect, patent ductus arteriosus, or systemic venous anomalies.[15,17,19] Systemic arterial collaterals occur in approximately 50% of patients with the adult form, but very few patients in this group receive substantial blood flow to the right lung from these vessels.[19] Consequently, unless an associated heart anomaly contributes to a left-to-right shunt, the increased ratio of pulmonary to systemic blood flow is caused by the anomalous pulmonary venous return. In such cases of the adult form, this shunt is typically less than 2:1, and the patients lead normal lives.

In contrast, patients with the so-called "infantile" form of scimitar syndrome typically present within the first few months of life with failure to thrive, cyanosis, respiratory distress, or congestive heart failure.[13-17,20] Sometimes the clinical clues are subtle, such as in an infant who is sick but exhibits only dextroversion on chest roentgenogram. At the other extreme, an infant may have a complex congenital heart defect, such as aortic arch hypoplasia, hypoplastic left heart syndrome, ventricular septal defects, pulmonary vein stenosis, or left ventricular outflow tract obstruction.[13-17] Systemic arterial flow to the right lung is more common in the infantile form than in the adult form. In fact, these systemic arteries supply the lower lobe and sometimes segments of the middle or upper lobes where pulmonary artery branches are lacking.[16,20,21]

Another characteristic of the infantile form (and often the factor responsible for the severe symptoms as well as the poor prognosis of this form) is pulmonary hypertension, which has been reported in 43–100% of patients with this form.[13,15-17,20] Factors thought to con-

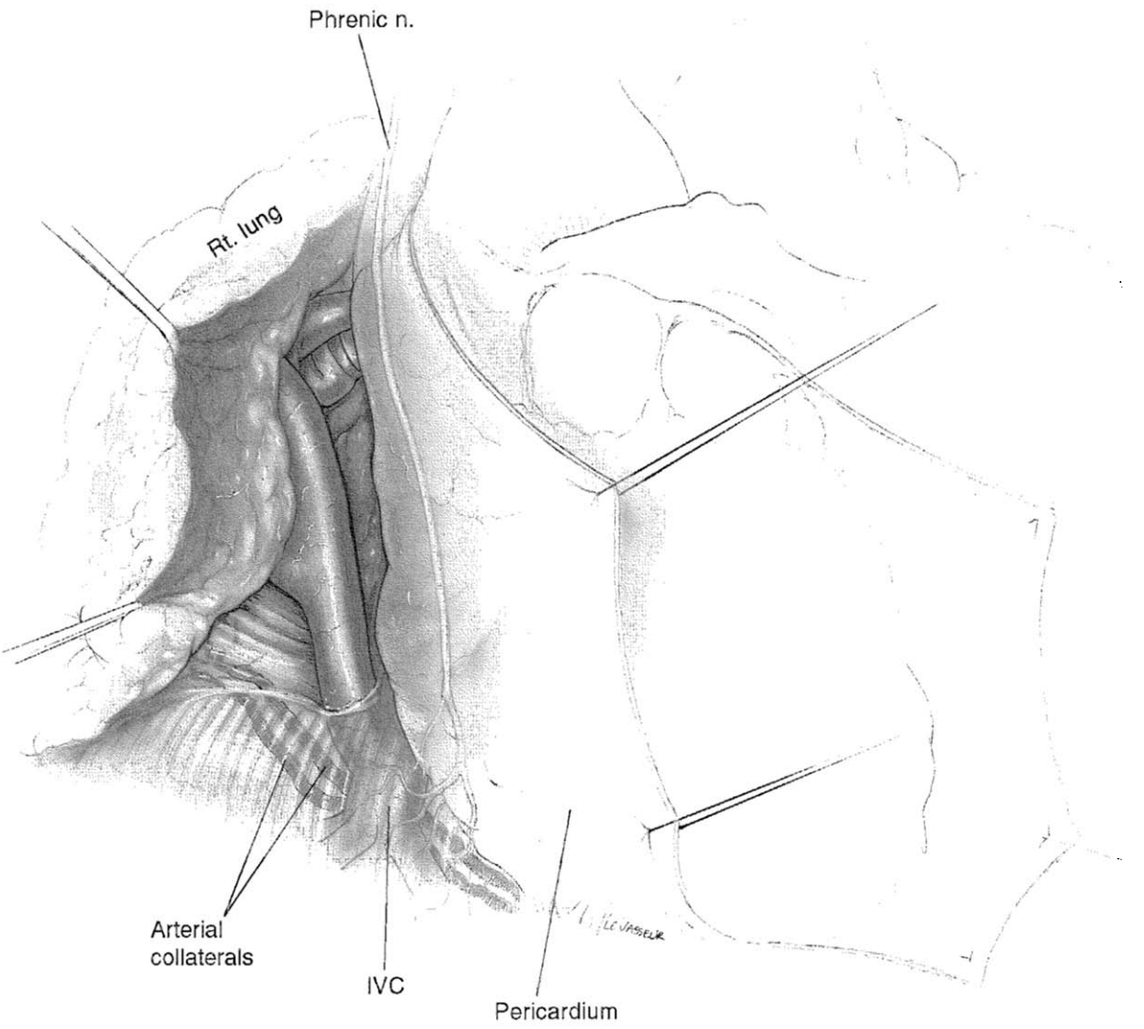
tribute to the pulmonary hypertension include (1) a left-to-right shunt from the anomalous pulmonary vein to the inferior vena cava; (2) stenosis of the scimitar vein or other pulmonary veins; (3) an associated intracardiac shunt, such as a ventricular septal defect; (4) reduction of the right pulmonary vascular bed due to lung hypoplasia; (5) failure of the pulmonary circulation to adapt postnatally in the face of excessive pulmonary blood flow[21]; and (6) a left-to-right shunt from the systemic arteries that supply the right lung to the pulmonary veins and ultimately to the inferior vena cava. Some authors propose that the systemic arterial shunt is the primary cause of the symptoms and the origin of the pulmonary hypertension in the infantile form.[20-22]

In any patient with suspected scimitar syndrome, both right and left heart catheterizations are needed to confirm the diagnosis. Angiography of the heart's right side will demonstrate the anomalous pulmonary vein and pulmonary artery anatomy, whereas aortography will delineate the systemic collateral supply to the right lung. Hemodynamic measurements will detect pulmonary hypertension if present, and hemoglobin saturation data are used to calculate shunt fractions. In a patient presenting with predominantly respiratory infection or dyspnea, bronchoscopy, computed tomography of the chest, bronchography, and ventilation scintigraphy may be used to evaluate the bronchial anatomy and estimate functional lung volume.

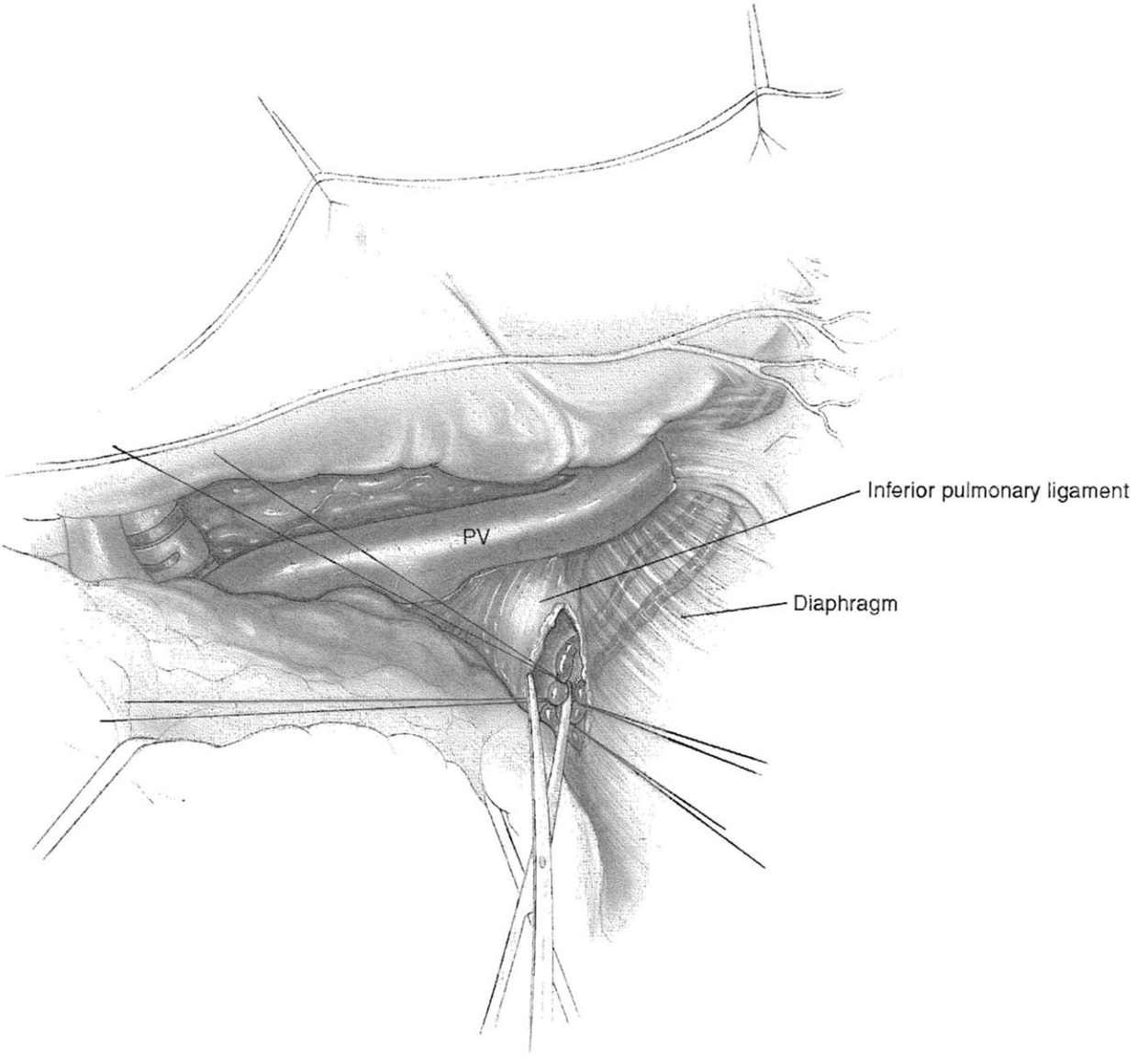
Currently, no single therapy is considered superior for treating scimitar syndrome. Because this anomaly is rare, prospective randomized evaluation would require a lengthy study and the participation of multiple centers. In addition, the long study duration would add other variables, such as changes in medical technology, patient care, and surgical expertise, which may impact significantly on outcome. The literature to date reports several small series each of which spans multiple decades, and it is from these data that we outline the therapeutic options for scimitar syndrome.

Most authors agree that systemic arterial collaterals should be eliminated to treat pulmonary hypertension and heart failure.[13,15-17,20-22] These vessels can be surgically ligated or embolized through catheter-based techniques. Any additional left-to-right shunt exceeding 2:1 should be corrected. However, preoperative studies should determine whether an associated cardiac defect, the anomalous pulmonary venous return, or both contribute significantly to the shunt. When the scimitar venous flow accounts for less than a 2:1 shunt by itself, surgical correction may be unnecessary. In contrast, when the left-to-right shunt through the scimitar vein exceeds 50%, this flow should be redirected into the left atrium by one of various techniques to lessen the volume load on the right ventricle and decrease the risk of late arrhythmias and exercise intolerance. Three options are available for such a repair: (1) The scimitar vein can be disconnected from the inferior vena cava and an anastomosis made between it and the left atrium directly or to the right atrium and the flow channled through the atrial septum by suturing an intra-atrial baffle; (2) the anomalous connection between the scimitar vein and the inferior vena cava can be left intact and a long baffle fashioned to redirect flow from this connection to the left atrium across the septum; or (3) the anomalous vein and the right atrium can be incised in parallel and then both openings sutured together to form a wide anastomosis. An intra-atrial baffle is then placed to guide flow from this anastomosis to the left atrium across an atrial septal defect. In some cases, a staged approach in which ligation or embolization of the systemic arterial collaterals is followed by surgical repair of the anomalous pulmonary venous return may be advantageous.[16,21,22] Lobectomy or pneumonectomy is best reserved for patients with recurrent infection, persistent hemoptysis, thrombosed intra-atrial baffles, or marked hypoplasia of the right lung.

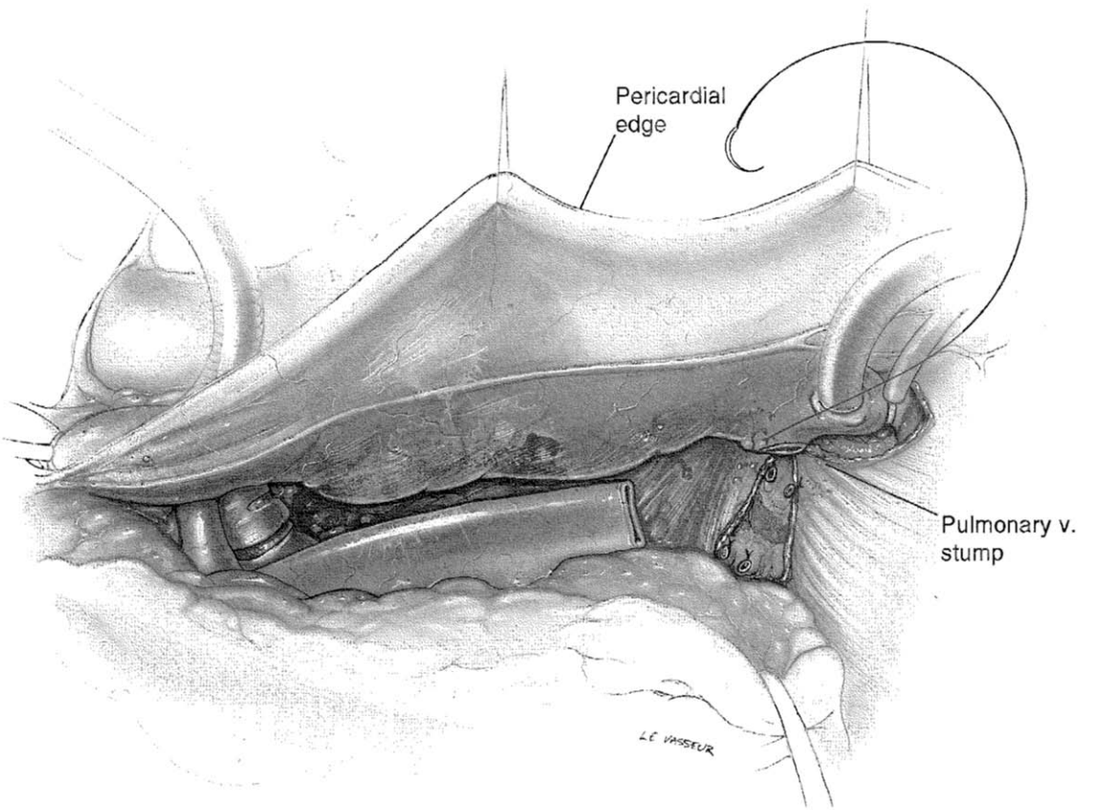
SURGICAL TECHNIQUE



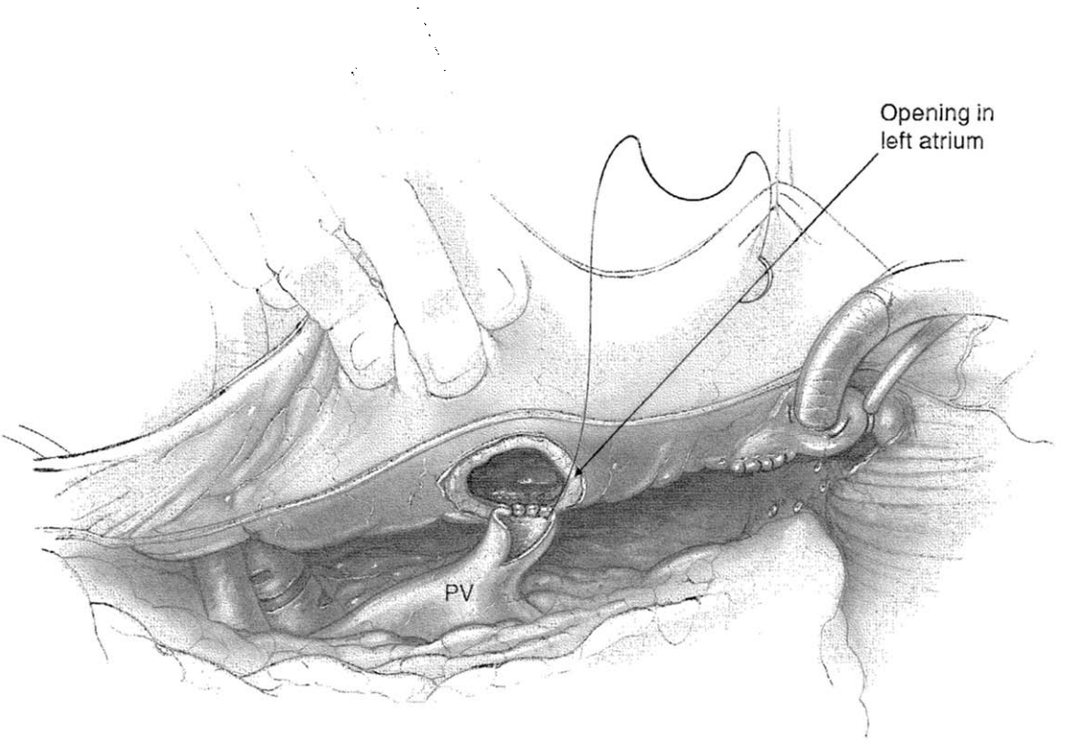
5 The chest is entered through a median sternotomy, the pericardium is entered, and the necessary great vessel dissection is done in preparation for aortic and bicaval cannulation. A pericardial cradle is created using stay sutures, with the ends of the sutures along the right side of the pericardium gathered in small clamps to allow freedom of movement of this pericardial edge. As shown here, the right pericardial edge is suspended and retracted to the patient's left. The right pleura is opened widely to expose the lung and hilum. A limited hilar dissection is done to free the anomalous pulmonary vein along its course from the lung to the inferior vena cava below the diaphragm. The diaphragm is incised from the inferior vena cava hiatus rightward and laterally to meet the point where the scimitar vein enters the vena cava. Note the diminutive size of the right lung and the systemic arterial collaterals as they travel within the peritoneum, pierce the diaphragm, and reach the inferior lung surface through the inferior pulmonary ligament.



6 Interruption of systemic arterial collaterals. The systemic arteries are dissected from the areolar tissue within the inferior pulmonary ligament and are divided between tied ends. The remainder of the inferior pulmonary ligament can then be cleared from the anomalous pulmonary vein to further mobilize it. Alternatively, the arteries can be simply ligated with titanium clips (Hemoclip; Weck Closure Systems, Research Triangle Park, NC).



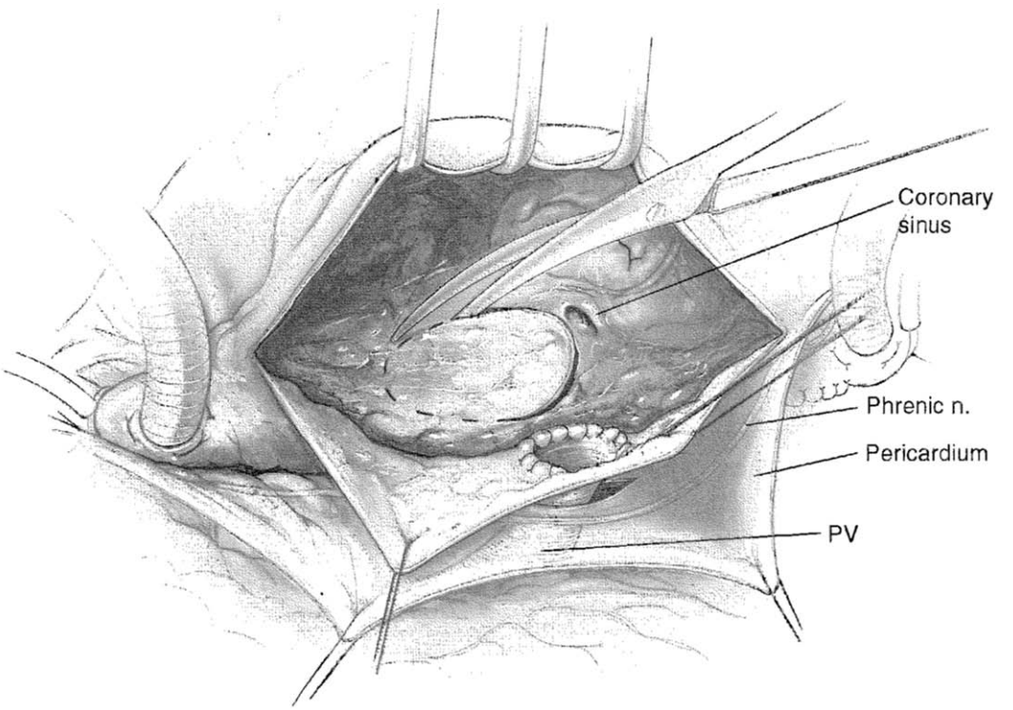
7 Division of the scimitar vein. The aorta is cannulated (Research Medical, Midvale, UT) and secured with a single purse-string suture (Ethibond; Ethicon, Somerville, NJ). Bicaval venous cannulation is done in a routine fashion with direct placement of the inferior vena cava cannula (DLP, Grand Rapids, MI) at the level of the hepatic vein entrance. Continuous cardiopulmonary bypass or deep hypothermic circulatory arrest can be used. The scimitar vein is divided at its junction with the inferior vena cava, and the caval stump is oversewn with a continuous running suture in two layers. The remainder of the vein is then mobilized to the hilum of the lung.



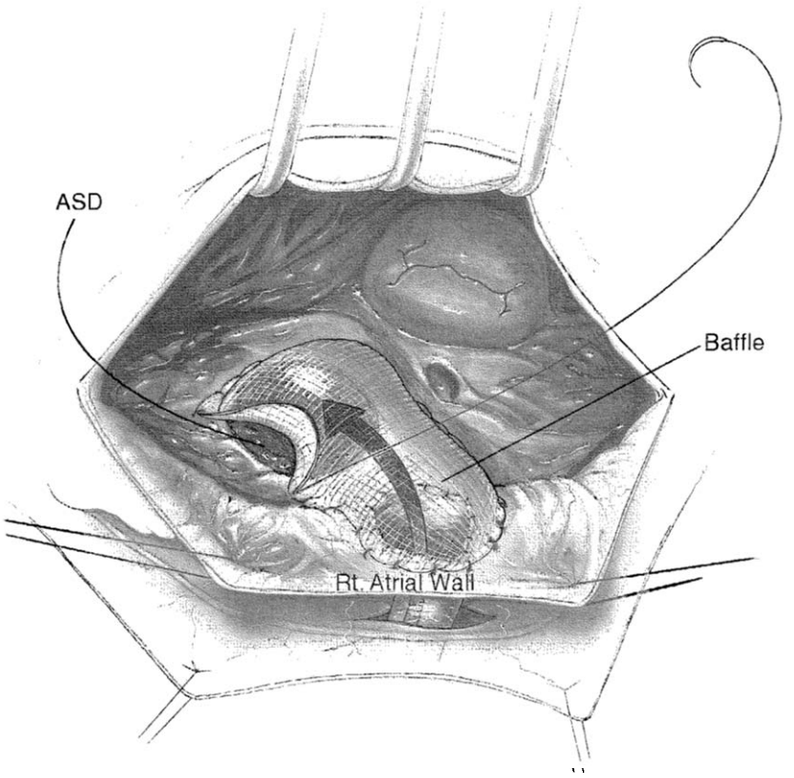
8 Creation of a venoatrial anastomosis. Several options are available for rerouting the right pulmonary venous return to the left atrium. One method is to create a direct connection between the anomalous pulmonary vein and the left atrium. This procedure can be done on cardiopulmonary bypass through a sternotomy, as shown, or can be performed through a left thoracotomy without bypass. The primary challenge in direct implantation is maintaining correct orientation of the vein so that it does not kink at the hilum when the lung is reinflated and the vein becomes more horizontal rather than vertical. Alternatively, an anastomosis can be fashioned between the pulmonary vein and the right atrium and the blood flow baffled across the atrial septum into the left side. The choice of the right or left atrium depends on the scimitar vein's length and mobility and on its relationship to the lung hilum. If the scimitar vein runs anterior to the hilum, then implanting it into the posterolateral wall of the right atrium is recommended. If the scimitar vein courses deep within the hilum, then implanting it into the posterior wall of the left atrium may be the best option. In either circumstance, the right pericardium is incised widely to create a window posterior to the phrenic nerve. The pulmonary vein can then be sized to the appropriate length to avoid redundancy or kinking. The orifice of the vein is enlarged by cutting longitudinally along its anterior length.

If the left atrium is chosen for the anastomosis, it is mobilized by dissecting from the interatrial groove posteriorly to adequately free the left atrial wall. A left atriotomy is then made in line with the course of the pulmonary vein as it passes through the window created in the pericardium. It is important to ensure that the vein lies correctly; distortion of this vessel can obstruct blood flow with consequent thrombosis. Enough atrial tissue should be resected to allow for a widely patent anastomosis. The pulmonary venoatrial anastomosis is constructed using a running continuous absorbable suture.

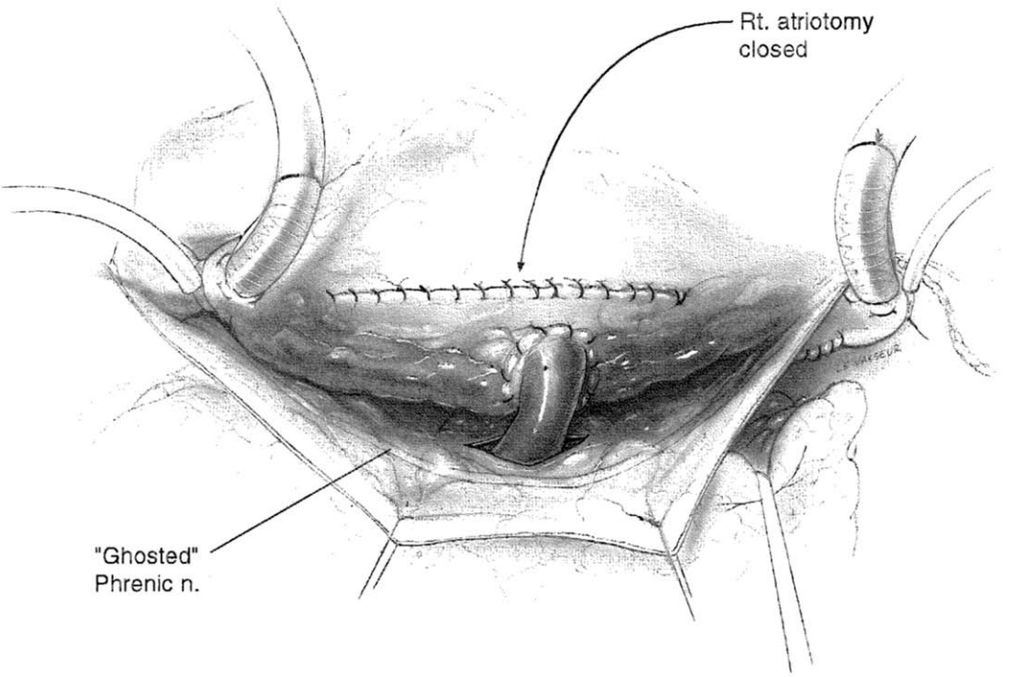
If the right atrium is chosen for the repair, then a large, vertical right atriotomy is made from the junction of the right atrium and inferior vena cava toward the appendage. From within the right atrium, the pulmonary vein is aligned with an existing or proposed site for an atrial septal defect. A right atriotomy is then made in the posterolateral wall while proper alignment of scimitar vein and atrial septal defect is maintained. The pulmonary venoatrial anastomosis is then constructed as described for the left side.



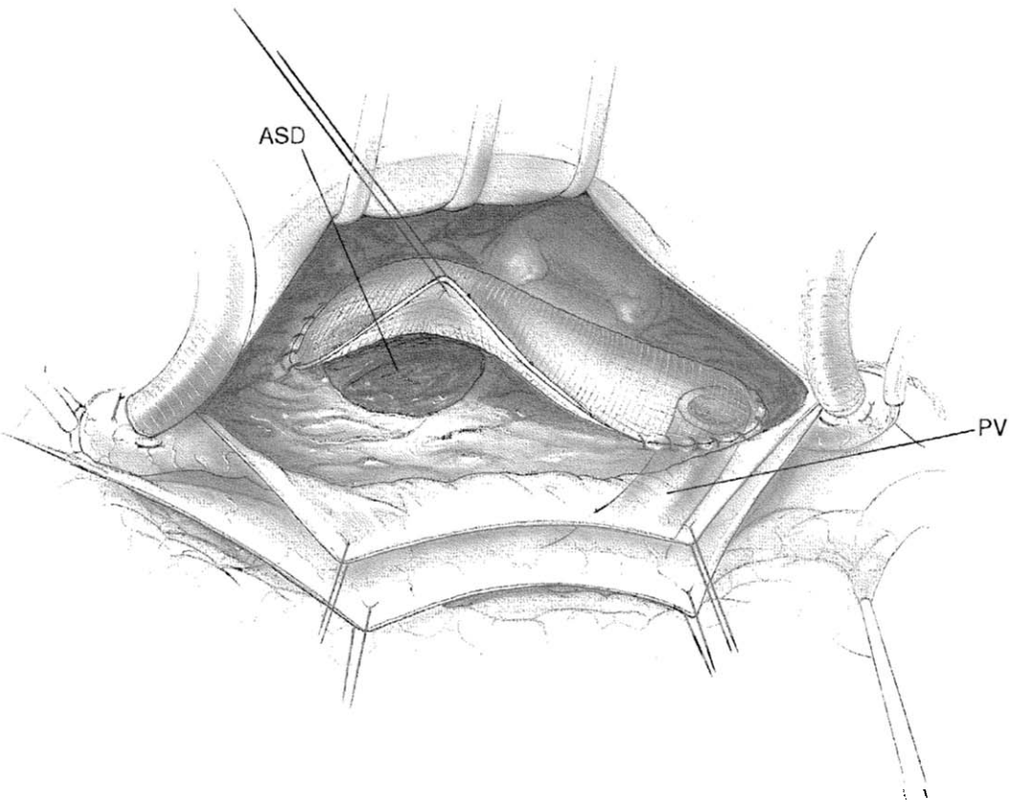
9 Creation of an atrial septal defect. If a secundum atrial septal defect is present, it must be large enough to accommodate all of the right pulmonary venous return without restriction. If it is not, then the defect must be enlarged by further excising septum until its diameter is at least as large as the diameter of the venoatrial anastomosis. When no defect exists naturally, the fossa ovalis should be excised liberally, as shown. An adequate rim of septal tissue should remain so that a baffle can be sewn in place without encroaching on the coronary sinus or the orifice of the superior vena cava.



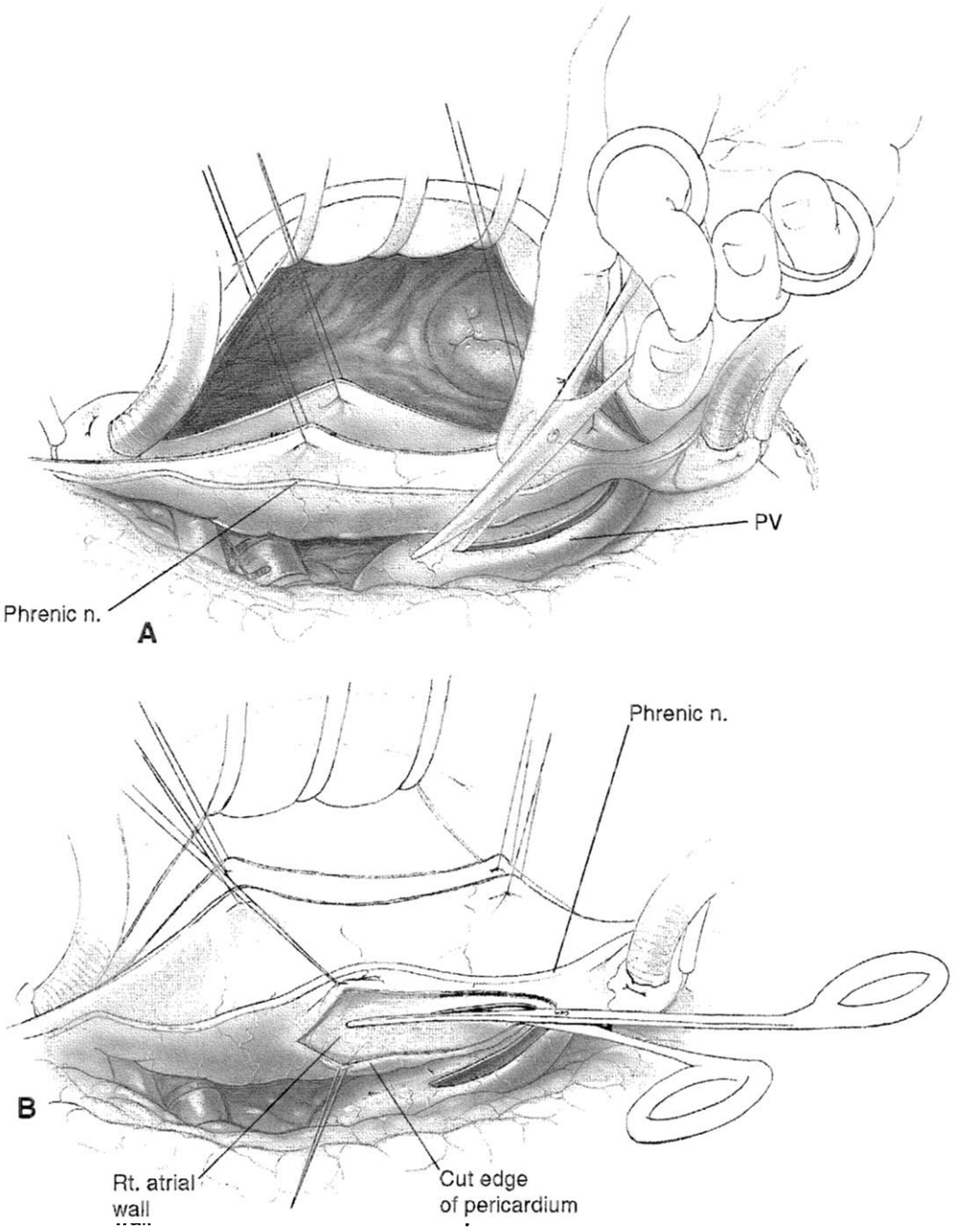
10 Baffle placement. A piece of native pericardium is harvested either when the initial pericardiotomy is done or when the baffle is constructed. Within the right atrium, the pericardium is cut to cover the orifices of both the venoatrial anastomosis and the atrial septal defect. The baffle must be cut with enough redundancy to allow it to billow as blood enters from the pulmonary vein en route to the left atrium. If the baffle is too taut, then flow across it will be restricted and thrombosis is possible. Excess fatty tissue is dissected from the pericardial surface, and this baffle material is positioned so the mesothelial surface faces the left atrial side. The baffle is then sewn into position using a continuous running suture. As mentioned previously, care must be taken not to narrow the ostia of the coronary sinus or the superior vena cava. The arrow indicates the direction of blood flow across the baffle.



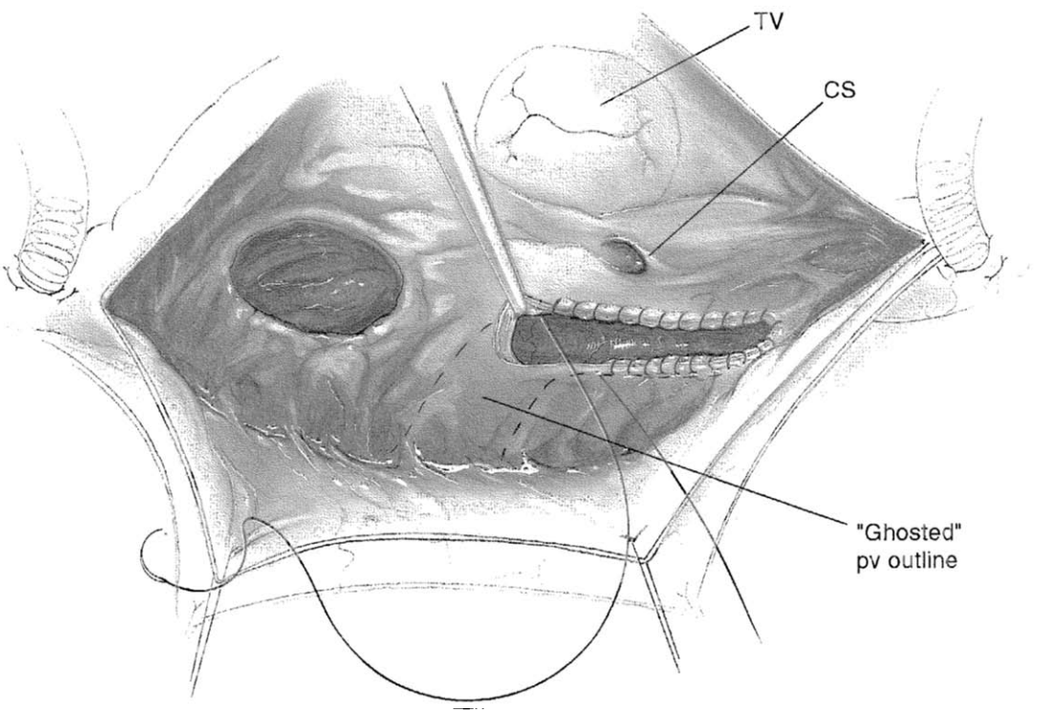
11 Closure of the right atriotomy. This figure shows a completed repair in which the anomalous pulmonary vein is connected to the posterolateral wall of the right atrium. Note the ghosted illustration of the phrenic nerve traveling uninterrupted along the pericardium.



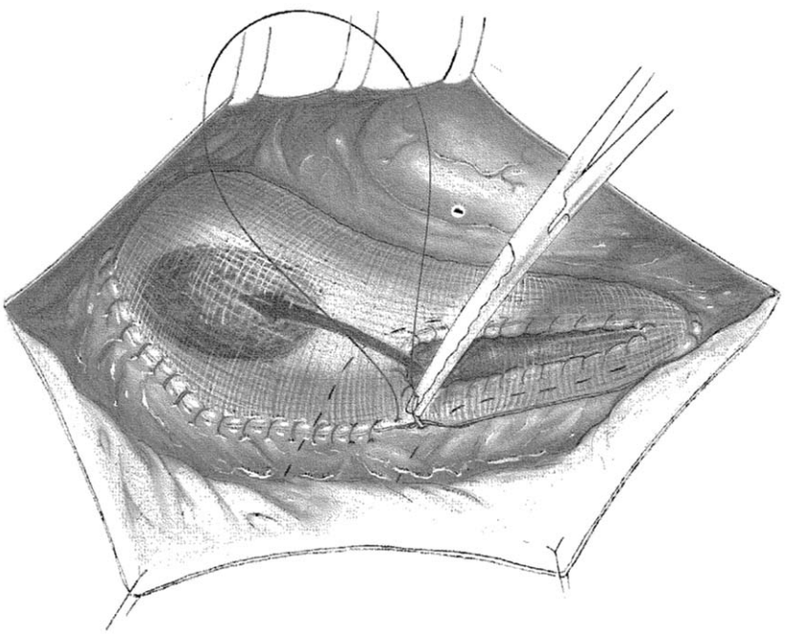
12 Long baffle repair. If the anomalous pulmonary vein courses along the posterior surface of the lung or appears too short to reach either atrium for a venoatrial anastomosis, then an alternative repair is creation of a long baffle from the vein's entry into the inferior vena cava up to an atrial septal defect. In this case the scimitar vein is not divided, and a vertical right atriotomy is created from the base of the atrial appendage to the proximal inferior vena cava just short of the venous cannula. As in the case of a pulmonary vein–to–right atrium anastomosis, if an atrial septal defect is small or not present, then one must be enlarged or created to allow unlimited flow from right to left. Native pericardium is used for the baffle, which is oblong-shaped and must be of adequate width to accommodate the entire right pulmonary venous return without restriction. The baffle is sewn into position with a continuous running suture, with care taken to avoid compromising coronary sinus or superior vena caval return.



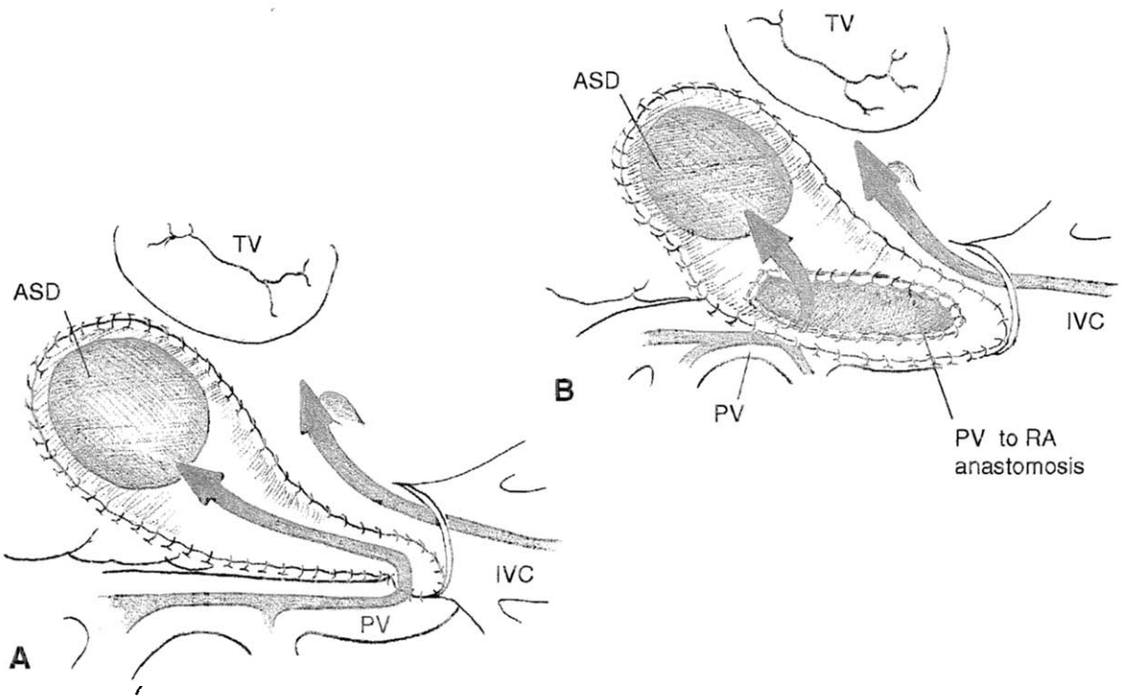
13 (A) Side-to-side venoatrial anastomosis. Another alternative to repairing the anomalous right pulmonary venous drainage is to incise both the posterolateral wall of the right atrium and the medial wall of the scimitar vein, and then join them in a widely patent anastomosis. The standard vertical right atriotomy is made and extended onto the inferior vena cava just short of the venous cannula. The pericardium is incised posterior to the phrenic nerve starting at its reflection onto the inferior vena cava and extending superiorly above the right lung hilum. Beginning at the inferior vena cava, the scimitar vein is cut along its medial length from its orifice to the confluence of its branches at the hilum. (B) The right atrium is incised. Beginning at the inferior vena cava from the orifice of the scimitar vein, the right atrium is cut vertically along its posterolateral wall for a length equal to that of the incision in the vein.



14 Creation of the anastomosis. From within the right atrium, the scimitar vein and the posterolateral right atriotomy are aligned so that the most posterior sides and the most anterior sides of both incisions are apposed. The pulmonary vein is then sewn to the right atrium using a continuous running absorbable suture.



15 Baffle placement. As with the previous baffle repairs, the atrial septal defect must be large enough to allow unrestricted right pulmonary venous flow into the left atrium. Native pericardium is cut to a size that will cover the pulmonary venoatrial anastomosis and the atrial septal defect, yet be redundant enough to accommodate right lung venous return without restriction. The baffle is sewn into position using a continuous running suture, with care taken to not narrow the orifices of the coronary sinus or the superior vena cava. The right atriotomy is then closed.



16 Schematic illustrations comparing the long baffle technique and side-to-side pulmonary venoatrial anastomosis for repair of scimitar syndrome. (A) In long baffle repair, pulmonary venous return must flow directly inferiorly to the orifice of the scimitar vein and then change abruptly to a superior direction, making nearly a 180° turnabout. This baffle technique, particularly in a low-pressure pulmonary venous circuit, may produce stenosis, promote stasis, and lead to thrombosis. (B) In a side-to-side pulmonary venoatrial anastomosis, pulmonary venous return flows directly across the baffle to the atrial septal defect in nearly a horizontal plane. This flow pattern is possible because the scimitar vein is opened and the anastomosis is widely patent. Theoretically, this technique reduces the risk for stenosis and obstruction of inferior vena caval return.

Results

The results for treating scimitar syndrome must be evaluated while keeping several factors in mind. First, the infantile form of the disease is more severe than the adult form. Second, most studies are small and retrospective, and thus conclusions about optimal therapy are based in part on chance outcomes, not necessarily outcomes resulting from a specific intervention. Third, because this syndrome is rare, all studies have been compiled over several decades. As such, these studies do not account for changes in surgical technique and in the care of critically ill patients that have evolved over the study periods. But despite these limitations, useful information obtained by many investigators forms the basis for treatment recommendations for scimitar syndrome.

In the infantile form of scimitar syndrome, medical management typically carries a mortality of 33–100%.[13,15,16,20] Conversely, a single study from Najm and colleagues[17] showed only one of seven infant deaths in those not undergoing surgery or catheter-based intervention. However, whether these seven infants had pulmonary hypertension or heart failure is unclear from this study. This information is important based on data from Dupuis and associates,[20] who reported on 25 patients with the infantile form of scimitar syndrome. Ten of these patients were treated medically, and mortality was 70%. These authors reported that the severity of symptoms, level of pulmonary pressure, and a younger age of presentation all influenced prognosis.

The surgical approach to the infantile form of scimitar syndrome has varied. Occlusion of the systemic arterial collaterals alone, either by surgical ligation or catheter embolization, is associated with poor results, with a mortality of 16–60%.[13,15-17,20] The long baffle repair has been associated with stenosis[17] or occlusion,[13] and subsequent pneumonectomy is often necessary. Division of the scimitar vein and reimplantation into either the left atrium or the right atrium with construction of an intra-atrial baffle has been done with possibly improved results. Huddleston and coworkers[13] reported that five patients treated with this technique were alive and well at midterm follow-up. However, only three patients required no further treatment. One patient underwent pneumonectomy for baffle occlusion, and another received a lung transplant for bilateral pulmonary vein stenosis.

Medical management for the adult form of scimitar syndrome has produced good results. Dupuis and colleagues[19] reported on 122 patients, 85 of whom did not undergo surgery; 79 of these 85 were alive and well. Two patients had chronic bronchitis and four had recurrent hemoptysis, but all six were able to conduct normal lives. In comparison, the 37 patients who underwent surgical repair, involving division of the scimitar vein and reimplantation into the left atrium, had mixed results. Twelve patients (33%) had a good outcome; 21 suffered postoperative respiratory complications and ultimately had poor long-term results. In addition, there was an 11% operative mortality not seen in the medically managed patients. The poorer outcomes for many surgically treated patients were attributed to thrombosis of the venoatrial anastomosis. Schramel[12] noted a similar result in a smaller series.

Interestingly, the long baffle technique has produced better results in the adult form than in the infant form. However, it has shortcomings. Murphy and associates[23] reported that five of six patients had a good short-term outcome, but the sixth patient had an immediate postoperative death. Similarly, in the series from Najm,[17] two of 11 (18%) adult patients who underwent long baffle repair developed pulmonary vein stenosis. One of these patients had the baffle excised, the atrial septal defect closed, and the scimitar vein left intact.

Regardless of whether the long baffle technique or division and reimplantation of the scimitar vein into an atrium is used, pulmonary venous stenosis, obstruction, and thrombosis remain the major obstacles to a successful repair in scimitar syndrome. As an alternative repair, we propose a side-to-side venoatrial anastomosis (Figs 13–15). This technique provides a widely patent anastomosis and allows for a short baffle. Compared to the long baffle repair, in which pulmonary venous blood flow must course through a sharp angle and be redirected nearly 180°, the side-to-side anastomosis allows for direct flow across the baffle to the left atrium on a horizontal plane (Fig 16). Compared to division of the scimitar vein and creation of an end-to-side venoatrial anastomosis, the side-to-side connection is larger and potentially less prone to hemodynamically significant stenosis. Although this side-to-side anastomosis has not been tested in a formal study, it theoretically will reduce anastomotic stenosis, stasis of blood flow, and pulmonary venous thrombosis.

Conclusions

Scimitar syndrome is a complex anomaly comprising various vascular, bronchial, and parenchymal derangements of the right lung. The clinical phenotypes are the infantile and adult forms. The infantile form is characterized by heart failure, pulmonary hypertension, associated cardiac defects, and a presentation before age 1 year. Therapy is best directed at correcting all malformations early. Occlusion of the arterial collaterals can be done as part of a complete surgical repair or by catheter embolization to temporize very ill infants. Associated heart malformations, particularly left-sided obstructing lesions or others that contribute

to pulmonary hypertension, should be corrected surgically along with repair of the anomalous pulmonary venous drainage. Medical management is ineffective in treating the infantile form.

The adult form is characterized by few or no symptoms or an incidental presentation later in life. Surgical intervention is not necessary unless the left-to-right shunt from the anomalous pulmonary vein exceeds 50% or the pulmonary vascular resistance is elevated without another identifiable cause.

In all cases, pulmonary resection should be limited to patients with thrombosed intraatrial baffles, chronic pulmonary infections, recurrent hemoptysis, or marked right lung hypoplasia.

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